Children's Eye Screening in the Primary Care Office: Rationale and Methods

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EDITOR'S NOTE: Due to space and color constraints, Figures 1-4 & 7 of this article are not printed in the journal. These figures are available online at dcmsonline.org. In the text, online figures are denoted with an asterisk (*).

Abstract: *Children's eye screening is essential for reducing vision loss in young children. The primary care physician plays an important role in eye screening. The screening exam elements are based on the child's age and are readily incorporated into routine medical examinations.*

Introduction

The primary care physician (PCP) plays a critical role in identifying vision loss among young children. Children's eye screening is widely recognized as a valuable public health service and is recommended as a standard component of pediatric primary care medicine by such organizations as the American Academy of Pediatrics, the American Academy of Ophthalmology, the American Academy of Family Physicians, and the U.S. Department of Health and Human Service. [1-6] Currently in the United States, however, many children fail to receive timely screening examinations. [6,7] The most efficacious approach to reducing vision loss in children is to increase the prevalence of eye screening, and primary care involvement is essential for this to occur. New educational initiatives and new technologies are being developed to improve the accuracy and frequency of screening examinations, but adequate tools already exist.

The Case for Children's Eye Screening

Many eye diseases of early childhood are not apparent without appropriate testing. In contrast to adults experiencing vision loss, young children with unilateral and even significant bilateral vision loss will tend not to complain. The target conditions for childhood eye screening are related to the age of the child. Likewise, the screening tests performed are dependent on the age of the child.

Target Conditions

In the first years, the primary targets for screening are cataract and retinoblastoma**;** blinding conditions that are more successfully treated with early detection.[8] Screening for these conditions is primarily based on detection of the red reflex in each eye. An abnormal whitish pupillary reflex color is termed leukocoria. Although the combined incidence of retinoblastoma and infantile cataract is low, the consequences of missing these diagnoses may be serious.

As the child becomes older, the primary screening target becomes amblyopia, a form of visual impairment affecting 3% to 4% of children. [9,10] In the process of screening for these conditions, signs of other eye abnormalities may also be detected.

Retinoblastoma is the most common malignant intraocular tumor of childhood and the seventh most common pediatric malignancy, with an incidence of 1:15,000 to 20,000 live births. [11] Each year in the United States 200 to 300 infants are diagnosed. Most cases are diagnosed by 3 years of age. Fewer than 25% of cases present with a family history of retinoblastoma. Most retinoblastomas are diagnosed after a child's relative or pediatrician notes leukocoria. Occasionally these white or yellow pupils are noted in photographs. Red reflex testing, a very quick test, should be repeated at each routine health visit. The second most common presenting sign of retinoblastoma is strabismus or malalignment of the eyes, and, therefore, every child with strabismus should be carefully checked for the presence of a normal red reflex in each eye. Unfortunately, many children with retinoblastoma or infantile cataract experience delays in detection or referral for ophthalmologic evaluation. [11-13]

Abramson *et. al.* reported that over 90% of leukocoria was initially detected by a family member of friend. [11] Parents may delay telling their pediatrician that leukocoria has been seen, and some pediatricians delay referral to an ophthalmologist of these seemingly asymptomatic children. [12] Mortality from retinoblastoma is related to the extent of disease, and, therefore, dependent on the tumor stage at diagnosis. The prognosis for preserving the eye and preserving useful vision is also related to the extent of disease. With the recent successes of chemoreduction and focal treatment, there are several modalities available for treating smaller retinoblastomas without enucleation. [14,15] *(*Figure 1)* When a retinoblastoma is first detected by a pediatrician's red reflex test rather than observations by the patient's family, there may be a greater chance of preserving the eye. [13]

Although most commonly an adult-onset condition, **cataracts** may be congenital or develop in infancy. The incidence of infantile cataracts is estimated at between 1 and 6 per 10,000 live births. [16] Treatment of a visually significant infantile cataract involves surgery followed by postoperative optical correction with glasses, a contact lens or intraocular lens implant. Permanent vision loss in cases of infantile cataract is usually due to visual deprivation amblyopia. [17] Timing of treatment is critical, as cataracts rapidly cause sensory deprivation amblyopia. Before the value of early treatment was recognized, vision results following treatment for unilateral infantile cataracts were almost uniformly poor.

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In the past two decades, excellent results have been achieved with treatment in the first few weeks of life. [17-19] This places new pressure on the primary care physician to detect cataracts in early infancy.

Amblyopia is a loss of vision caused by deficient visual stimulation of one or occasionally both eyes during the years of visual development. Several ocular conditions may interfere with normal development of vision and, thereby, cause amblyopia. Amblyopia is most often caused by strabismus. Anisometropia, a difference in refractive power between the two eyes, causes amblyopia without any noticeable change in a child's appearance. Visual deprivation, for instance from a cataract or a ptotic eyelid, can cause severe amblyopia and require early intervention. Although amblyopia is caused by ocular conditions, the defect of amblyopia develops within the brain. [20] With time, untreated amblyopia leads to permanent vision loss. [21]

Strabismus is a condition of ocular misalignment. In strabismus, the visual axes of the two eyes are not directed toward the same object. Strabismus is classified by the direction of the deviating eye, as in esotropia, exotropia, and hypertropia. *(*Figure 2)* Subtle expressions of strabismus are often diagnosed later than more obvious cases, and, therefore, the angle of strabismic deviation correlates inversely with the density of amblyopia. [22] The corneal light reflex test, the cover test, and the Bruckner test are used to detect strabismus.

Justifying Amblyopia Screening

According to the World Health Organization, conditions appropriate for mass screening should be highly prevalent in the population, should be treatable at the time of detection, and should result in significant impairment to the individual without timely treatment. [23] Amblyopia meets these criteria. Amblyopia is certainly prevalent, with roughly one amblyopic child per kindergarten class. Although usually treated successfully when detected in the preschool years, amblyopia is the leading cause of monocular vision loss among adults in the United States. [24] The failure of the prior generation to minimize vision loss from amblyopia is multifactorial, related to a poorer understanding of amblyopia, limited vision screening efforts and difficulties with treatment compliance. Today, unfortunately, not all of these factors have been eliminated. Delay in treatment leads to worsened visual outcomes. [21] Uncorrected amblyopia may harm school performance and the ability to learn and may later cause problems with adult self-image. [25] The presence of amblyopia places the opposite eye at increased risk for vision loss. [26] Also, ophthalmologists may be more reluctant to operate on an eye with partial vision loss when the opposite eye is amblyopic. [27]

Studies from Sweden and Israel have demonstrated that the incidence of amblyopia can be significantly reduced in the general population by screening for amblyopia by four years of age. [28,29] A vision screening pilot project in the United States was not as successful, however, because of variability in testability and referral rates. [30] Amblyopia treatment outcomes are better with early detection and treatment, due to faster improvement in visual acuity and better compliance with treatment. [31-34] The Agency for Healthcare Research and Quality's U.S. Preventive Services Task Force (USPSTF) highly rates vision screening at age 3 to 4 years for cost effectiveness and clinically preventable burden. [3] Compared to other ophthalmologic interventions, the cost of identifying and treating amblyopia is low, and successful treatment results in decades of improved vision. [35,36]

Government Recommendations

The Maternal and Child Health Bureau recommends preschool vision screening but recognizes that additional work is required before the ideal screening strategy is determined. [23,30,37] The national health objectives of the U.S. Department of Health and Human Services (Healthy People 2010) include increasing the proportion of preschool children who receive vision screening. Their 2002 National Health Interview Survey found that 36.3% of children less than 6 years old had ever undergone vision testing. [6] Florida Medicaid's Child Health Check-Up program (formerly EPSDT) mandates standardized vision screening for children 3 years old and older. [38]

The Case for Primary Care Screening

Sweden and other countries have developed national systems to detect amblyopia in the preschool age group. In Sweden, more than 90% of children are screened by age 4 years, resulting in significantly less amblyopia in the population. [29] The prevalence of preschool vision screening is lower in the U.S. Nationally, only 21 to 36% of children younger than 6 years have ever had their vision tested. [6,39] Charity-based pre-school screening programs in the United States report even lower penetrance of 10% and 14% in selected statewide screening programs. [40] Of the 55% of pediatricians who responded to a recent survey, 35% reported screening 3-year-old children using eye charts, 73% 4-year-old children, and 66% 5-year-old children. [41] These results are similar to those of a 1992 study. [42] These studies may overestimate the actual number of screening exams performed.

The Vision Is Priceless Council (VIP Council), a Northeast Florida charitable organization, performs the majority of charity-based preschool vision screening in this region. Within Duval County, for the 2005-2006 academic year, the VIP Council screened 4,257 3-year-olds (39% of the population), 6,400 4-year-olds (58%), and 3,685 5-year-olds (33%). Despite this great effort, it is clear that many children are missed by daycare and preschool-based screening. Through their contact with children in the Medical Home and their respect among parents, pediatricians and family physicians are well positioned to increase the prevalence of screening, diagnosis, and treatment of amblyopia. To capture the most preschool children, a U.S. vision screening model should include preschool/daycare-based screening as well as primary care office screening for amblyopia.

Screening itself, of course, does not improve vision. Follow-up confirmatory examination and treatment are the necessary next steps. Despite the efforts of preschools, daycares, and screening organizations, many parents do not follow up with

a comprehensive eye exam, perhaps not trusting the results of the screening exam. [9] Parents' trust in the primary care physician may help convey the importance of a follow-up comprehensive eye exam. There is limited published data related to the efficacy of vision screening in the primary care setting. [30,43] For screening infants and nonverbal children for cataract and retinoblastoma, there is no charity-based safety net, and screening is dependent on the primary care physician's exam.

Barriers to Primary Care Preschool Vision Screening

The primary care physician faces several obstacles to enacting universal children's eye screening. For preschool vision screening, the ideal test does not yet exist. Difficulties exist with test reliability, time required to test, cost and reimbursement, and training. The success of the Swedish screening system, however, suggests that low-tech pre-literate eye chart screening can be effective. Screening with pre-literate eye charts requires staff training and time, however. Screening a 3-year-old child with the Lea Symbols chart may take 3 or more minutes. Screening a 4-year-old, however, usually requires less than 2 minutes. The vast majority of 3-year-olds (92%), 4-year-olds (97%), and 5-year-olds (98%) can be screened with the Lea Symbols chart in daycare and preschool settings. [44]

Barriers to office screening reported by pediatricians include lack of cooperation by children (49%), screening being too time-consuming (23%) and lack of training (15%). [41] In response to difficulties in eye chart screening, particularly in children age 3 years and younger, newer screening technology has been developed. Photorefraction and autorefraction devices such as the MTI Photoscreener, the SureSight Vision Screener and the Retinomax Autorefractor are designed to detect abnormalities of ocular refractive power and/or binocular alignment that are precursors of amblyopia. These devices offer the advantage of rapid objective testing that requires minimal patient cooperation. Problems with the devices include their relatively high cost and the high rates of false-positive results. [43] The Vision in Preschoolers Study found that in the hands of eye care professionals, the SureSight and Retinomax autorefractors were comparable in accuracy to the Lea Symbols eye chart. [45] Other studies have shown success with photorefraction devices in mass preschool screening programs. [46] Currently less than 10% of pediatric practices use these automated devices. [41] The most recent American Academy of Pediatrics (AAP) photoscreening policy statement indicates the potential advantages of photoscreening without fully endorsing the techniques. [47]

Inadequate reimbursement is also a barrier to primary care preschool vision screening. [35] The 99173 CPT code ("screening tests of visual acuity, quantitative, bilateral ") is not consistently used by pediatricians. One limited study reported that 51% of pediatricians performing preschool vision screening billed this CPT code and that most who used the code received some reimbursement. [41] For screening of children ages birth to 3 years, tests such as the red reflex test are rapid and easily incorporated into a pediatric examination, and adoption should be more responsive to education initiatives.

Practice guidelines alone are likely to have insufficient impact on physician behavior. [48] Training in preschool vision screening is not a standard part of pediatrics and family medicine residency programs. [49] Phillips et. al. defined a physician's recognition of a problem but failure to act as "clinical inertia." Clinical inertia is more likely to be present in the management or detection of asymptomatic conditions, particularly when there is a lack of education, training, and practice organization related to the condition. [50] One-time office-based training in vision screening has been shown to improve screening rates among 3-year-old children, but not dramatically. [51] As a next step in learning how to improve primary care vision screening rates, an interactive CD, Pediatric Vision Screening, was recently distributed to pediatricians and family physicians in Florida. This interactive CD, created by Nemours and the VIP Council, reviews the target conditions of screening and demonstrates recommended vision screening techniques. An attached survey will be used to measure opinions and practices related to children's eye screening among primary care physicians. Ultimately, education and practical training in preschool vision screening during pediatrics and family medicine residency may be more effective than efforts directed toward practicing physicians. [49,50]

Children's Eye Screening Tests

When to Screen: The appropriate screening tests to be performed are dependent primarily on the child's age. [4] From birth and at each well child exam until reading an eye chart, red reflex testing is performed. The external appearance of the eyes and eyelids is also inspected. From 3 months through 5 years, corneal light reflex testing is performed. Corneal light reflex testing may be combined with red reflex testing in the Bruckner test. An assessment of fixation and following behavior is performed beginning at two to three months of age. From 6 months to 5 years, cover testing is performed. From 3 to five years, pre-literate eye chart testing is performed. This eye examination strategy is intended for use in apparently normal children without any condition or family history placing them at increased risk for eye disease. Other recommendations apply for certain children at greater risk for eye disorders. For example, the AAP recommends an ophthalmology referral for all children with Down syndrome by 6 months of age. [52] Because of an increased risk of strabismus, amblyopia, and refractive errors, children born prematurely should undergo follow-up ophthalmologic examinations even after complete resolution of retinopathy of prematurity. [53] A family history of conditions such as retinoblastoma and congenital cataract should also prompt ophthalmology referral.

Red Reflex Test: The red reflex is produced by light reflecting from the retina. Anything obstructing a view of the retina, such as a cataract, will block the red reflex. An abnormality of the retina, such as a retinoblastoma, may also eliminate or distort the red reflex. To perform the test,

a direct ophthalmoscope is used to view each pupil, 12 to 18 inches from the child. If a cataract is present, the red reflex may be absent, dulled or patchy. *(*Figure 3)* Alternatively, the pupil may appear white or yellow. An eye harboring a retinoblastoma may have a white or a shiny "cat's eye" pupillary reflex. *(*Figure 4)*

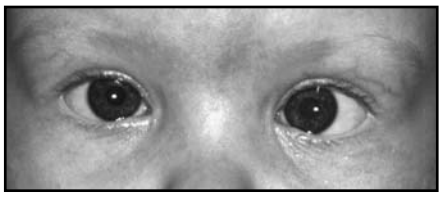
External Inspection of the Eyes: In addition to looking for the red reflex in an infant, the external appearance of the eyes and eyelids should be observed. Eyelid ptosis may cause amblyopia or be a sign of a neurologic abnormality. Both small and large corneas are associated with eye disease. A large cornea is a presenting sign of infantile glaucoma. Asymmetry of pupil size, anisocoria, may be the result of eye disease or a neurological abnormality.

Fixation Behavior Assessment: While performing eye tests on preverbal children, assessing the child's fixation behavior is also recommended to qualitatively evaluate a young child's vision. Visual behavior in infancy evolves rapidly. By six weeks of age, the child should make some visual response to one's face. By two months, there should be some following behavior demonstrated. If poor fixation and following are noted binocularly after three months of age, an ophthalmology referral is recommended. By four to six months, fixation behavior may be tested monocularly, with each eye alternately covered. If poor fixation and following occur with one eye occluded, or if the child strongly protests having one of his or her eyes occluded, the uncovered eye may have poor vision.

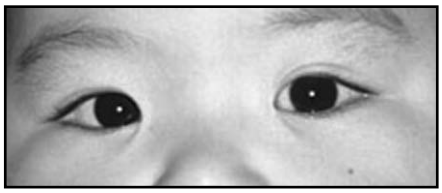
Corneal Light Reflex: The smooth convex surface of the cornea causes a sharp reflection from any point source of light such as a penlight or direct ophthalmoscope. The position of this light reflex, relative to the underlying iris and pupil, is dependent on the alignment of the eye. To perform the corneal light reflex test, one directs a light on the child's eyes and observes the corneal light reflexes of both eyes simultaneously. A temporal location of the light suggests esotropia, an inward misalignment of the eye. A nasal location of the light reflex suggests exotropia, an outward misalignment of the eye. *(Figures 5, 6)*

Figures 5 & 6 Corneal Light Reflex Test

(Right, top) Estropia is demonstrated by the temporal location of the left corneal light reflex.



(Right, bottom) Normal position of the corneal light reflexes in pseudoesotropia.

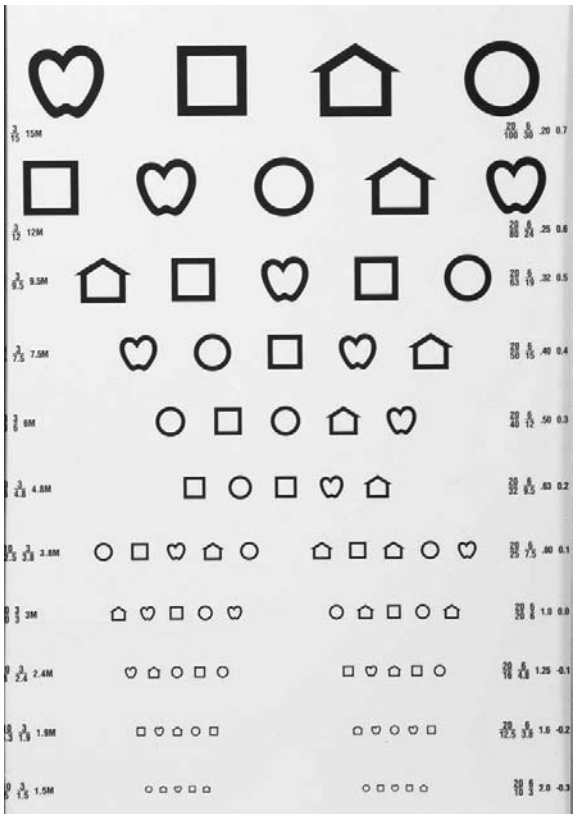


The Bruckner Test: The Bruckner test is a combined, simultaneous corneal light reflex and red reflex test, performed with the direct ophthalmoscope. The Bruckner test may reveal strabismus, anisometropia, leukocoria, and anisocoria. A direct ophthalmoscope is held 2 to 3 feet from the child. The red reflexes and the corneal light reflexes of both eyes are viewed simultaneously. Interpretation of the corneal light reflex is the same as with the standard corneal light reflex test. As with other red reflex testing, an abnormal red reflex in the Bruckner test may indicate cataract or retinoblastoma. In addition, a brighter red reflex in one eye may indicate that the eye is strabismic. A difference in red reflex color may also indicate anisometropia, a difference in optical power between the eyes and a risk factor for amblyopia. *(*Figure 7)* Any abnormality of corneal light reflexes, red reflex symmetry or pupil symmetry should prompt an ophthalmology referral. [54]

Cover Tests: Cover tests are used to assess eye alignment and diagnose strabismus. In the cover test, each eye is sequentially occluded as any resultant eye movement of the opposite is observed. A re-fixation movement may indicate the presence of strabismus.

Pre-literate Eye Charts: The primary goal of eye chart screening in the preschool age is to detect amblyopia. Preliterate eye charts are available for screening children not yet ready for screening with letters. The HOTV Chart and the Lea Symbols Chart are recommended for screening three- and four-year-old children. *(Figure 8. See dcmsonline.org for full version)* Testability is improved by testing children at ten feet from the eye chart.

Figure 8 Lea Symbols Chart (Useful ages 3 & up)



For the Lea symbols chart, three and four year old children must read the majority of symbols on the 20/40 line of the chart. For five-year-olds, the critical line may be advanced to 20/32. If either eye does not reach this level, the patient should be referred for further evaluation.

Conclusion

By performing timely vision screening exams, primary care physicians play a critical role in preserving the vision of young children. Retinoblastoma, infantile cataracts, strabismus and amblyopia are not always obvious to parents or pediatricians. Treatment success improves with early detection. Early detection and intervention are key to providing the best opportunity for excellent vision in every child. These four disorders are the primary targets of vision screening in young children. Early detection of these conditions leads to better long-term outcomes.

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